Neonatal Cardiac Distress

A Practical Approach to Recognition, Diagnosis, and Management

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■ *The death rate among neonates with cardiovascular disease is 50 percent during the first six months, with the majority dying during the first month. With early diagnosis most of these babies could be saved. In approaching the diagnosis of cardiac distress in the newborn, it is important to remember that the types of cardiovascular disease which cause symptoms and death early in life are quite different from those in older children. Lesions such as hypoplasia of the left heart, transposition of the great arteries, endocardial fibroelastosis, pulmonary atresia, mitral atresia, tricuspid atresia and truncus arteriosus are common, not rare, causes of cardiac distress in the newborn.*

A classification of neonatal cardiovascular diseases into seven pathophysiological groups is presented as a basis for an effective, practical approach to the differential diagnosis of the potentially lethal lesions. This approach is simplified further since over 90 percent of babies with cardiac distress have one of three lesions: (1) Large left-to-right shunt (characterized by the presence of massive plethora on the chest roentgenogram), (2) Large right-to-left shunt (association with intense cyanosis) or (3) Severe obstruction (including hypoplasia of the left heart, which is the most common cause of death due to cardiac distress during the first week of life).

THE EARLY RECOGNITION of cardiovascular disease in neonates is difficult but extremely important since the natural attrition rate is 50 percent during the first six months, with most deaths occurring during the first month.[31] If there is early recognition of the problem, most of these babies can be salvaged by aggressive diagnostic and surgical management.[13]

When should a serious cardiovascular lesion be suspected, how is the differential diagnosis made and what can be done for potentially lethal cardiac disease in the neonate? The basis of a practical approach to these questions is the classification of neonatal heart disease into seven lethal lesions as listed in Table 1. This classification stresses the physiological, not the anatomical, abnormality and is more helpful to the clinician since the physiology of these lesions is more pertinent to the clinical findings and course.[15]

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TABLE 1.—*Neonatal Cardiac Distress—The Seven Lethal Lesions*

Lesion	*Diagnostic Tip Off*	*Differential Diagnosis*		*Treatment*
		Acyanotic	*Cyanotic*	
1. LEFT-TO-RIGHT SHUNT	X-ray (plethora)	ASD, VSD, PDA, AV Communis, AP Window, AV Fistula	SV, Truncus, Incomplete Trans., *TA* (type C), *TAPVD**	Division or repair Band Septoplasty Transplantation
2. RIGHT-TO-LEFT SHUNT	Cyanosis		Trans., TOF, *TA*, *PS*, PA, *Ebstein's*	Septectomy Shunt Valvuloplasty Glenn
3. OBSTRUCTION	Electrocardiogram (hypertrophy)			
a. Without systemic hypoperfusion		Coarc., AS, MS, *PS*	*HLH* (Ao Atresia), MA, *TAPVD* below diaphragm, *TA*	Aortoplasty Valvuloplasty Septectomy
b. With systemic hypoperfusion	Pulses (weak or absent)	Hypoplastic Ao without ductus	*HLH* without ductus	?Shunt + Band
4. PRIMARY MYOCARDIAL DISEASE	X-ray (cardiac enlargement) No murmur Epigastric impulse	EFE, Myocarditis, Ab. Lt. Cor., Glycogen Storage, Cor. vas., Idio. Hyper., Vent. Diverticulum, (Pericardial Dis.)		Digitalis Ligation Excision
5. REGURGITATION	Murmur	MI, AI, PI		?Valvuloplasty ?Replacement
6. AIRWAY OBSTRUCTION	Stridor	Vascular ring		Division
7. ARRHYTHMIAS	Electrocardiogram	PAT (WPW), Flutter, Block (Corr. trans.)	*(Ebstein's)*	Digitalis Cardioversion Pacemaker

*The lesions in italic appear under more than one category.

LEGEND

Ab. Lt. Cor.=Aberrant left coronary artery; AI=Aortic insufficiency; Ao=Aorta; AP Window=Aortopulmonary window; AS=Aortic stenosis; ASD=Atrial septal defect; AV Communis=Atrioventricularis communis; AV Fistula=Arteriovenous fistula; Coarc.=Coarctation of the aorta; Corr. Trans.=Corrected transposition; Cor. Vas.=Coronary vasculitis; EFE=Endocardial fibroelastosis; Idio. Hyper.=Idiopathic hypertrophy; MI=Mitral insufficiency; MS=Mitral stenosis; PAT=Paroxysmal atrial tachycardia; PDA=Patent ductus arteriosus; PI=Pulmonary insufficiency; PS=Pulmonary stenosis; SV=Single ventricle; TA=Tricuspid atresia; TAPVD=Total anomalous pulmonary venous drainage; TOF=Tetralogy of Fallot; Trans.=Transposition of the great arteries; Vent.=Ventricular; VSD=Ventricular septal defect; WPW=Wolff-Parkinson-White syndrome.

When Should a Lesion be Suspected?

Suspicion of a cardiovascular lesion in the newborn is difficult since the signs and symptoms are usually occult.[10] Additionally, contrary to common belief, the majority of neonates with serious lesions do not have murmurs and the detection of a cardiac problem therefore depends more on inspection and palpation than on auscultation.[11] Whenever heart disease is suspected, a chest roentgenogram and an electrocardiogram should be performed and these may provide the only definite clue to the presence of a serious problem. When the physician suspects a significant cardiac lesion, pediatric cardiology consultation is indicated. In view of the limitations of physical diagnosis and because the prognosis depends on exact diagnosis, immediate or early cardiac catheterization and angiography are often advisable.[4] The important clinical, roentgenographic and electrocardiographic findings are summarized below.

Clinical Findings

Detection of pronounced cyanosis is simple and obvious; however, evaluation of minimal cyanosis can be misleading in the normally polycythemic neonate. Episodic cyanosis is almost always due to pulmonary or central nervous system disease rather than cardiac disease. The cyanotic baby who "pinks up" when placed in high tension oxygen usually has pulmonary disease but this change can also occur in cardiac babies who have interference with gaseous exchange due to congestion from left-sided decompensation. Babies with minimal right-to-left shunts have a "rosy" appearance which the unwary physician may interpret as a sign of robust good health.

Differentiation of cardiac cyanosis from cyanosis due to central nervous system disease or pulmonary disease is often dependent on the presence of other signs and symptoms of heart disease. Differential cyanosis is pathognomonic of preductal coarctation with or without transposition.[19] Chest deformity due to cardiac enlargement is rare in the neonate and is usually due to aortic atresia or a cardiac tumor. Tachypnea (resting respiratory rate greater than 40) is the earliest sign of left-sided decompensation and is usually accompanied by intercostal and sternal retractions.

Pulmonary edema should be suspected in all neonates with respiratory distress, and each should be examined carefully for other signs of cardiac disease. Airway obstruction due to a vascular ring must be suspected in babies with stridor, particularly when the stridor is intensified during nursing.

Evaluation by palpation of the peripheral pulse volume and the precordial cardiac impulse is very important but is often neglected in the newborn. The classic weak or absent femoral pulses of coarctation are well known but seldom sought. A sudden generalized decrease in pulse volume is characteristic of vasoconstriction of the ductus in a baby with hypoplasia of the left heart (HLH). Significant hemodynamic overloading is best assessed clinically by palpation of the precordial impulse. Normally, the neonatal precordial impulse is quiet and tapping and located at the xyphoid angle. In the neonate, location of the maximal cardiac impulse at the apex is pathognomonic of a left ventricular overload. A hyperdynamic impulse at the apex or xyphoid angle indicates a diastolic or volume overload (left-to-right shunt). Precordial thrills are common with ventricular septal defect, pulmonary stenosis and aortic stenosis.

Hepatomegaly is the most common sign of right-sided failure. The congested liver edge is blunt, tender and points anteriorly. A presystolic hepatic pulsation ("a" wave) is found when there is obstruction to right atrial egress as with tricuspid atresia or severe pulmonary stenosis.

Dependent edema is a rare and late sign of right-sided failure in the neonate.

As previously stated, heart murmurs are infrequent in neonates with cardiac distress, and also, contrary to common belief, the intensity of murmurs does not correlate well with the severity of disease. Although often difficult to detect, gallop rhythm is almost always present in neonates in early congestive failure.

Chest roentgenography

Roentgen examination of the neonate should usually be limited to a portable anterior posterior examination since these babies are usually in acute distress and cannot safely be moved. Lateral, oblique and barium views are seldom helpful anyway. Of paramount importance is an appreciation of the decided limitations of chest roentgenographic examination in the newborn.

The clinical findings (including the electrocardiogram) must be considered for optimal roentgen interpretation (Figure 1 and Table 2). Interpretation should include evaluation of: (1) Heart size and position, (2) Pulmonary vascular mark-

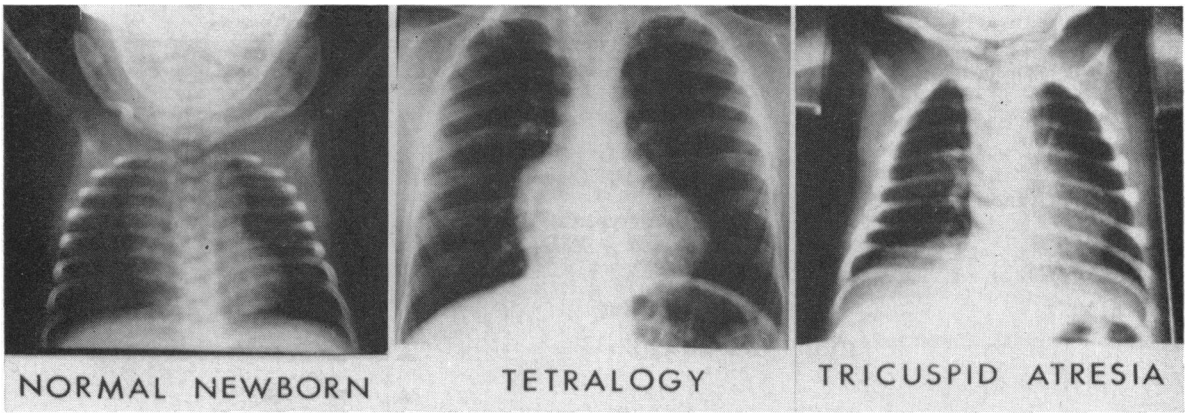

Figure 1.—Chest roentgenograms of a normal newborn, and newborns with tetralogy of Fallot and tricuspid atresia. Roentgenographic differential diagnosis without knowledge of the clinical and electrocardiographic findings is usually of limited value in the neonate.

ings, (3) Configuration of the main pulmonary artery and (4) Size and position of the aortic arch.

Cardiac enlargement should be suspected whenever the cardiothoracic ratio exceeds 60 percent; however, accurate assessment is complicated by the presence of the thymic shadow (Figure 2) and variations in heart size associated with respirations (Figure 3). In the frontal projection, right ventricular enlargement usually is associated with elevation of the cardiac apex *(cor en sabot)* and left ventricular enlargement with downward pointing of the cardiac apex. Usually the newborn with cardiac distress has generalized cardiac dilatation, and attempts to evaluate specific chamber enlargement are fruitless.

Dextrocardia and isolated levocardia are important clues to the probable presence of severe cardiac anomalies (Figure 4). Although errors are frequent, the pulmonary vascularity is classified into one of three groups — increased, normal or decreased. Both active and passive congestion can be mimicked or masked by pulmonary inflammatory disease or the respiratory distress syndrome.

Accurate roentgen or fluoroscopic differentiation of active from passive pulmonary congestion is not possible in the neonate and Kerley's B lines are rarely seen. The shadow of the main pulmonary artery is usually of increased convexity in patients with large left-to-right shunts, valvar pulmonary stenosis, pulmonary hypertension and incomplete transposition of the great vessels of the Taussig-Bing[25] or OBGVRV* types and is concaved in tetralogy and complete transposition.

The size, shape and position of the aortic arch are important in the diagnosis of certain lesions including isolated pulmonary stenosis (the aortic arch is almost always on the left), tetralogy of Fallot (the aortic arch is on the right in 34 percent of cases), coarctation of the aorta, aortic stenosis and vascular ring.

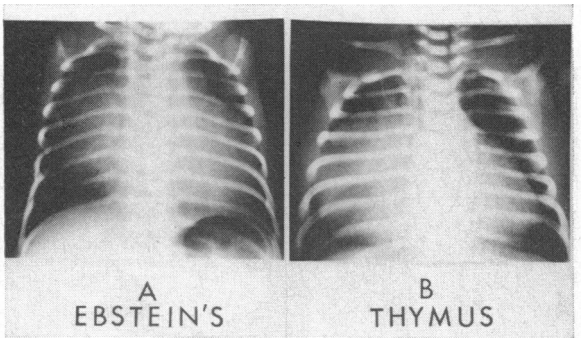

Figure 2.—Enlargement of thymus can mimic cardiac enlargement. Both of the chest roentgenograms illustrated are from two-day-old babies. The patient whose x-ray film is on the left had Ebstein's anomaly and died at five days of age. The apparent enlargement of the cardiac silhouette of the roentgenogram on the right is due to thymic, not cardiac, enlargement which had totally regressed by six months of age.

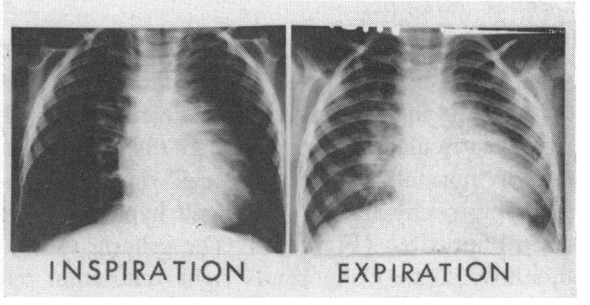

Figure 3.—The chest roentgenograms illustrated are both from the same patient who had "pink" tetralogy of Fallot and were taken within ten minutes of each other. The exposure on the left was taken during full inspiration and the exposure on the right during expiration.

*Origin of both great vessels from the right ventricle.

TABLE 2.—*Radiographic Diagnosis of Cardiovascular Disease in the Neonate*

Clinical Findings		EKG Findings	Diagnosis	CTD	MPA	Ao	Comments
PVM ▲ *	Cyanosis	RVH	HLH, MA	▲	►	▼	
			Trans.	▲	▼	►	"Eggshape"
			TAPVD	▲	▲	▼	"Snowman"
			TAPVD (below diaph.)	►	►	▼	
		LVH	TA (type C)	▲	►	►	
			HLH + VSD	▲	►	►	
		CVH†	SV	▲	▲	►	
			Truncus	▲	▼	▲	Ao. arch rt. (40%)[22]
	No cyanosis	RVH	ASD	▲	▲	►	Ao. arch rt.
		LVH	PDA	▲	►	▲	
			AP Window	▲	►	▲	
			AV Fist.	▲	►	►	
		CVH	VSD	▲	►	►	
			AV Com.	▲	►	►	
			AV Block	▲	►	►	
PVM ▼	Cyanosis	RVH	TOF	►	▼	▲	Ao. arch rt. (34%)[22]
			PS	▲	▲	►	Ao. arch lt.
		LVH	TA (type A)	►	▼	▲	Ao. arch rt. (10%)[22]
			PA	▲	►	►	
			Ebstein's	▲	►	▼	
PVM ►	No cyanosis	RVH	PI	▲	▲	►	
		LVH	Coarc.	▲	►	▲	
			AS	▲	►	▲	
			AI	▲	►	▲	
			MI, PMD	▲	►	►	LA ▲
		No VH	PAT	▲	►	►	
			Vas. Ring	►	►	►	Tracheal Compression[17]

*Active or passive.
†CVH or either RVH or LVH.

► Normal or no increase in size.
▲ Increased size or prominence.
▼ Diminished size or prominence.

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AI=Aortic insufficiency; Ao=Aorta or aortic; AS=Aortic stenosis; ASD=Atrial septal defect; AP Window=Aortopulmonary window; AV Block=Atrioventricular heart block (Complete); AV Com.=Atrioventricularis communis; AV Fist.=Arteriovenous fistula; Coarc.=Coarctation of the aorta; CTD=Cardiac transverse diameter; CVH=Combined ventricular hypertrophy; Diaph.=Diaphragm; HLH=Hypoplasia of the left heart; LA=Left atrium; LVH=Left ventricular hypertrophy; MA=Mitral atresia; MI=Mitral insufficiency; MPA=Main pulmonary artery; PA=Pulmonary atresia; PAT=Paroxysmal atrial tachycardia; PDA=Patent ductus arteriosus; PI=Pulmonary insufficiency; PMD=Primary myocardial disease; PS=Pulmonary stenosis; PVM=Pulmonary vascular markings; RVH=Right ventricular hypertrophy; SV=Single ventricle; TA=Tricuspid atresia; TAPVD=Total anomalous pulmonary venous drainage; TOF=Tetralogy of Fallot; Trans.=Transposition of the great arteries; Vas. Ring=Vascular ring; VH=Ventricular hypertrophy; VSD=Ventricular septal defect.

Electrocardiogram

The electrocardiogram frequently is of limited value in the neonate since most of the serious cardiac lesions in this age group are associated with right ventricular hypertrophy and the differentiation of "normal" or "physiological" right ventricular hypertrophy from "abnormal" hypertrophy is often impossible (Figure 5). The criteria used at this hospital for "abnormal" right ventricular hypertrophy are listed in Table 3. A "normal adult pattern" in the neonate is pathognomonic of left ventricular hypertrophy (Figure 6). As illustrated in Figure 7, the electrocardiogram is usually of value in differentiating dextroposition, dextrocar-

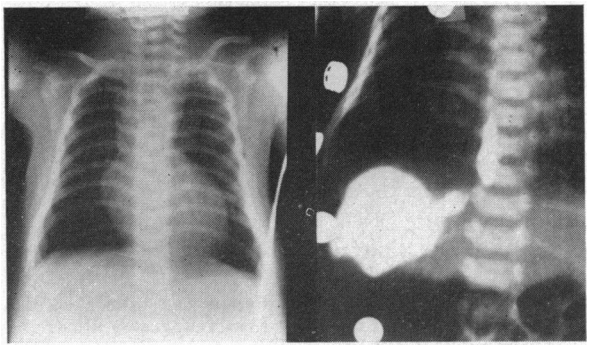

Figure 4. — Chest roentgenograms illustrating isolated levocardia. Notice that the gastric shadow is on the right while the cardiac shadow is on the left. The patient also had total anomalous pulmonary venous drainage below the diaphragm and tetralogy of Fallot.

TABLE 3.—*Electrocardiographic Criteria for "Abnormal" Right Ventricular Hypertrophy in the Neonate*

General Criteria	*Voltage Criteria*	*Intrinsicoid Deflection*
1. Frontal QRS axis > 180°	1. $R_{AVR} > 10$	1. Greater than 0.03 in V_1 (without IRBBB)
2. Pure R or qR in V_1	2. $R_{V_1} > 20$	
3. R/S ratio in $V_1 > 90\%$		
4. Upright T wave in V_1 beyond 48 hours		
5. Depressed ST segment and deeply inverted T wave in V_1		

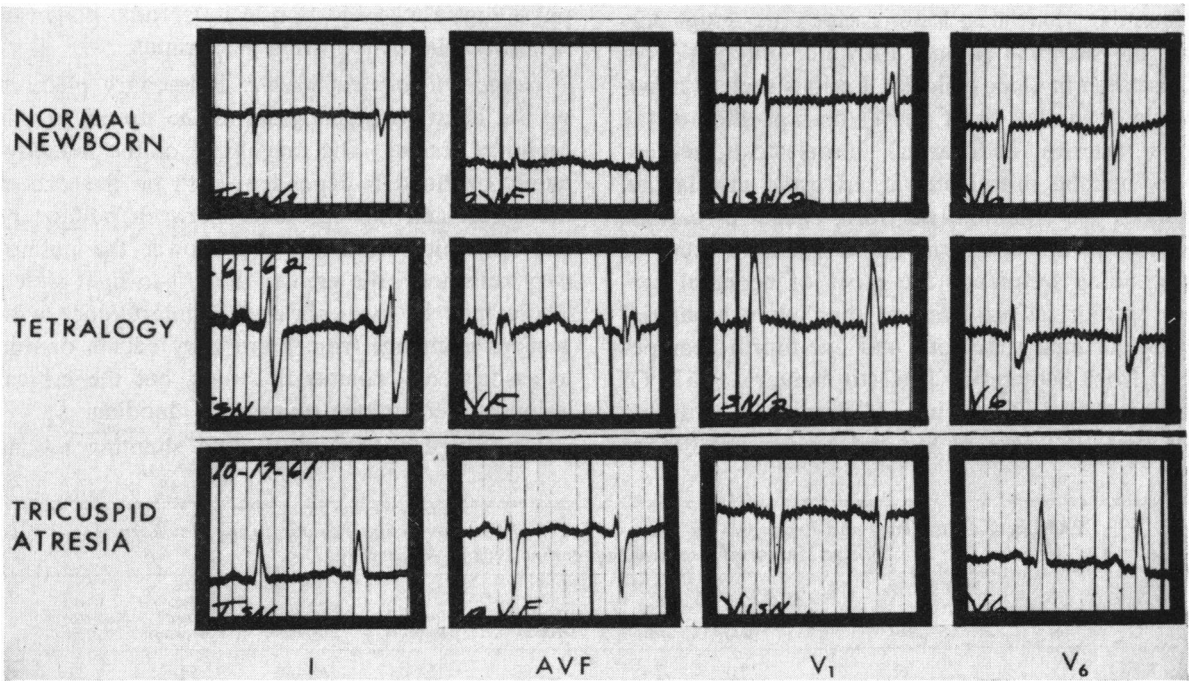

Figure 5.—The electrocardiogram of a normal newborn is compared with the tracing of a newborn with tetralogy of Fallot and a newborn with tricuspid atresia. The normal newborn tracing shows right ventricular dominance, similar to that of the newborn with tetralogy. The electrocardiogram of the neonate with tricuspid atresia is strikingly different showing left axis deviation and left ventricular dominance similar to an adult tracing.

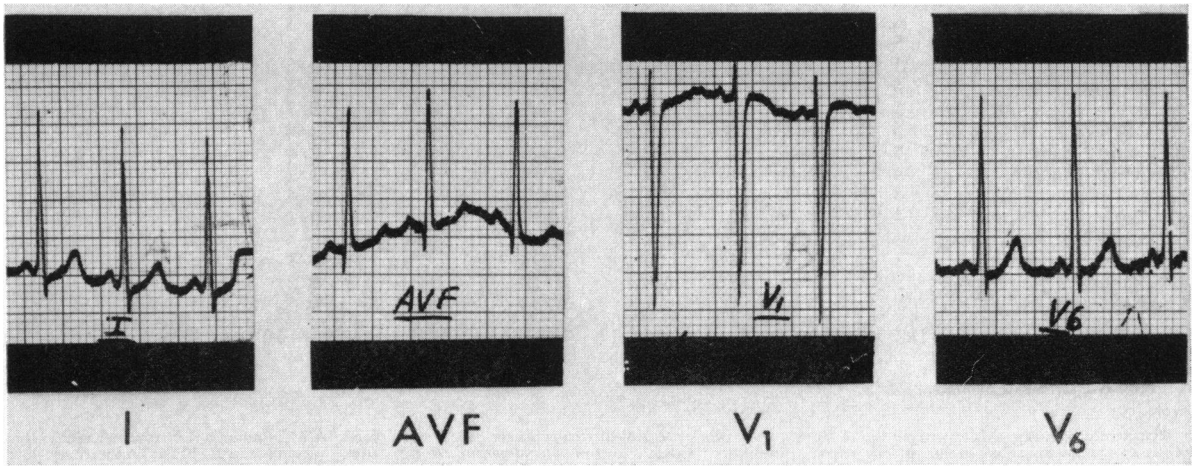

Figure 6.—A "normal adult" type electrocardiogram is always abnormal in a neonate. The tracing which is illustrated was obtained from a baby boy with single ventricle.

dia and dextroversion. The electrocardiogram is essential for the differential diagnosis of tachycardia, bradycardia and irregular rhythm and for the detection of abnormal depolarization patterns such as the Wolff-Parkinson-White syndrome (WPW).

Differential Diagnosis and What Can Be Done

In differential diagnosis it is important first to remember that heart lesions which are lethal during the neonatal period are quite different from those seen in older children. Lesions such as hypoplasia of the left heart (HLH), transposition of the great arteries, endocardial fibroelastosis, pulmonary atresia, mitral atresia, tricuspid atresia and truncus are common, not rare, causes of cardiac distress in the newborn. Table 4 lists in order of descending frequency the types of neonatal cardiovascular lesions seen at the Sutter Memorial Hospital during the four and one-fourth year period from September 1962 to January 1967. Of the total of 182 patients, 34 percent died during the first 12 weeks of life and 58 percent (35 of 61) of these deaths were due to the seven lesions listed above.

When using the classification of the seven lethal lesions (Table 1), the approach to the differential diagnosis is further simplified when it is remembered that over 90 percent of the causes of neonatal distress can be classified into only the first three pathophysiological groups: (1) Large left-to-right shunt, (2) Large right-to-left shunt and (3) Severe obstruction. The use of this pathophysiological classification in differential diagnosis is outlined in the following paragraphs.

Large left-to-right shunts. Pulmonary plethora on the chest roentgenogram is the tip-off to this group of lesions. The magnitude of the hemodynamic overload is dependent both on the size of the defect and the difference between pulmonary and systemic resistance — the lower the pulmonary resistance, the greater the left-to-right shunt. There may be cyanosis due to interference with gaseous exchange from pulmonary edema or due to associated venoarterial shunts, but the critical lesion is tremendous pulmonary flooding.

Uncomplicated left-to-right shunting lesions

TABLE 4.—*Incidence and Prognosis for Various Types of Cardiovascular Disease in the Newborn (Sutter Memorial Hospital September 1962 to January 1967)*

Lesions	No.	Age at Onset of Signs: 1st wk.	2-6 wks.	7-12 wks.	Cardiac Distress*	Surgical Survivors	Medical Survivors	Spontaneous "Cure" (47 cases)	Deaths (in first 12 wks.)
1. VSD	39	4	10	25	38%	3/3	36/36	51%	0
2. TOF	23	5	9	9	83%	3/4	14/19	....	6
3. PDA	15	5	4	6	74%	5/5	9/10	20%	1
4. PS	11	1	5	5	36%	1/1	10/10	....	0
5. HLH	7	6	1	0	100%	...	0/7	....	7
6. Trans.	7	3	4	0	100%	2/5	0/2	....	5
7. AS	6	2	1	3	50%	1/2	4/4	....	1
8. EFE	6	2	2	2	100%	...	3/6	33%	3
9. PA	6	4	2	0	100%	0/3	0/3	....	6
10. SV	6	0	5	1	83%	2/3	1/3	....	3
11. ASD	5	1	3	1	80%	1/2	3/3	40%	1
12. AV Block	5	3	1	1	20%	...	4/5	....	1
13. AV Com.	5	0	1	4	100%	1/2	1/3	20%	3
14. MA	5	3	2	0	100%	0/2	0/3	....	5
15. TAPVD	5	1	3	1	100%	0/1	3/4	....	2
16. Coarc., com.	4	0	2	2	100%	...	2/4	....	2
17. Coarc., uncom.	4	0	2	2	50%	1/2	2/2	....	1
18. PAT	4	1	2	1	100%	...	4/4	....	0
19. TA	4	1	2	1	100%	1/3	0/1	....	3
20. Vas. Ring	4	1	2	1	100%	4/4	...	....	0
21. Trans., incom.	3	2	1	0	100%	...	0/3	....	3
22. Truncus	3	1	2	0	100%	0/1	0/2	....	3
23. Misc.	5	2	2	1	100%	...	0/5	....	5
Totals	182	48	68	66	72%	25/43	96/139	15%	61 (34%)

*Congestive failure or hypoxia.

LEGEND

AS=Aortic stenosis; ASD=Atrial septal defect; AV Block=Atrioventricular heart block (Complete); AV Communis=Atrioventricularis communis; Coarc., com.=Coarctation of the aorta, complicated; Coarc., uncom.=Coarctation of the aorta, uncomplicated; EFE=Endocardial fibroelastosis; HLH=Hypoplasia of the left heart; MA=Mitral atresia; Misc.=Miscellaneous; PA=Pulmonary atresia; PAT=Paroxysmal atrial tachycardia; PDA=Patent ductus arteriosus; PS=Pulmonary stenosis; SV=Single ventricle; TA=Tricuspid atresia; TAPVD=Total anomalous pulmonary venous drainage; TOF=Tetralogy of Fallot; Trans.=Transposition of the great arteries; Trans., incom.=Transposition, incomplete; Vas. Ring=Vascular ring; VSD=Ventricular septal defect.

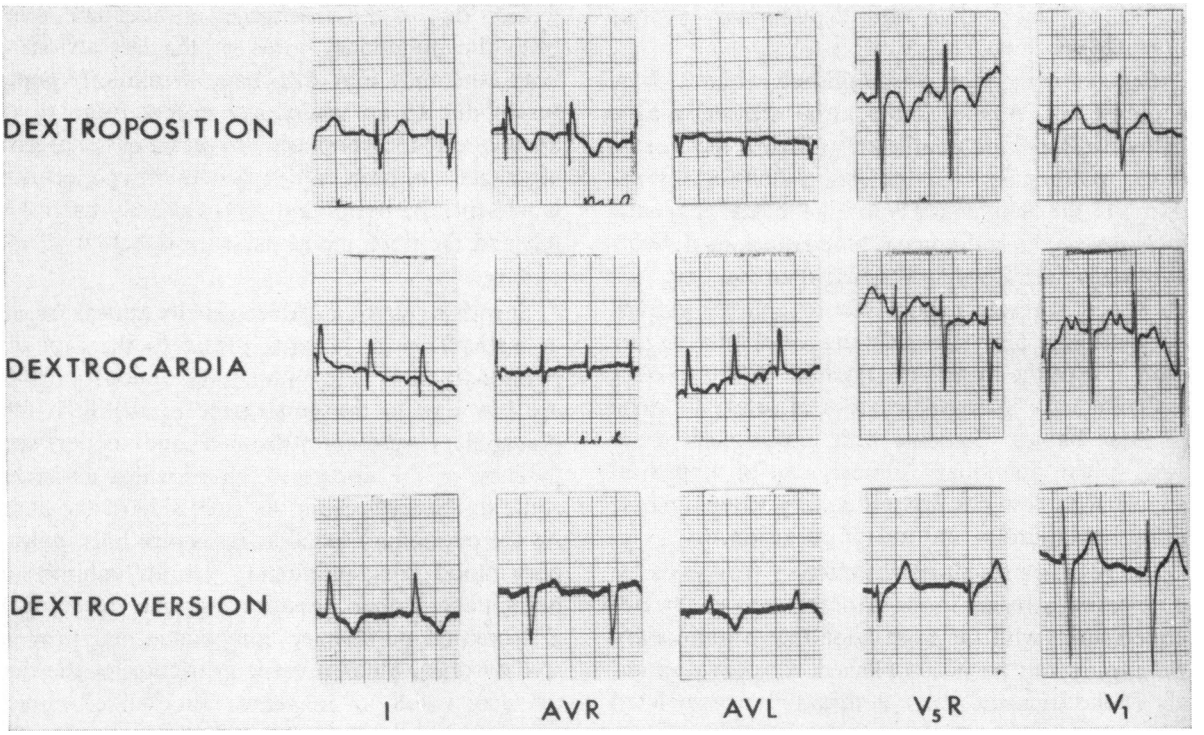

Figure 7.—Electrocardiographic differentiation of dextroposition, dextrocardia, and dextroversion is usually possible. The electrocardiogram in patients with dextroposition is normal except that the precordial transition zone is to the right. In mirror image dextrocardia both the P waves and T waves are inverted in Leads I and AVL and are upright in Lead AVR. In dextroversion only the T wave is inverted in Leads I and AVL and is upright in AVR.

usually do not cause significant symptoms until after the third week (Table 4). This late onset of distress is related to the relatively high pulmonary vascular resistance at birth. Conservative management is often indicated for patients in this category, particularly those with intracardiac shunts, due to the high neonatal surgical risk and the high percentage of lesions which undergo spontaneous closure (Table 4).

Since the clinical findings of ductus are usually atypical during the neonatal period and because the management and prognosis for ductus are so different from the other left-to-right shunting lesions, all patients in this group should be suspected to have ductus until proved not to by cardiac catheterization and angiography. These laboratory studies are therefore usually indicated early and often urgently in patients of this type.

Effective palliation can frequently be achieved by pulmonary artery banding in many of the other lesions associated with large left-to-right shunts[16] (ventricular septal defect, communis defect, aortopulmonary window, single ventricle, truncus, incomplete transposition, and tricuspid atresia type C[26]). Certain types of total anomalous pulmonary venous drainage can be corrected relatively easily by septoplasty.[40] The risk of corrective operation for the types of total anomalous pulmonary venous drainage which require transplantation of a common venous trunk is high and palliative atrial septectomy and pulmonary artery banding may be preferable.[5]

Neonatal congestive failure due to an uncomplicated atrial septal defect of the secundum type is rare. However, when there are progressive symptoms in spite of intensive medical therapy, open heart surgical correction of the defect is indicated and can be accomplished at relatively low risk.[34]

A large peripheral arteriovenous fistula, often intracranial or intrahepatic, can cause occult congestive failure and is occasionally diagnosed by the astute clinician who performs careful auscultation of the head and abdomen.[20]

Large right-to-left shunts. Sudden acute distress (hypercyanotic "spells") and severe metabolic acidosis due to anaerobic metabolism are common complications of these lesions.[44] Transposition of the great vessels is differentiated from the other types of severe cyanotic lesions by the presence of pulmonary plethora on the chest roentgenogram. Early atrial septectomy is mandatory for babies with transposition, not only to increase the arterial

saturation, but also to relieve pulmonary venous hypertension.[3]

Septectomy is best accomplished without thoracotomy by "popping" the atrial septum with a balloon (Rashkind) catheter[38] (Figure 8). A systemic pulmonary anastomosis (shunting procedure) of the Schumaker type[42] is the best approach to increase the pulmonary blood flow in patients with severe tetralogy and tricuspid atresia type A.[26] However, severely hypoplastic or aplastic pulmonary arteries may cause failure of shunts of any type. Use of the hyperbaric chamber seems to offer a significantly increased margin of safety in some of these babies.[2] Patients with uncomplicated severe valvar pulmonary stenosis can be improved at relatively low risk by pulmonary valvuloplasty, using hypothermia and inflow occlusion.[12]

There is at present no satisfactory treatment for pulmonary atresia. These patients are usually operated upon with the faint hope that a pulmonary valvotomy can be accomplished. Ebstein's anomaly of the tricuspid valve is usually not associated with hypoxia during the first few months of life. When there are progressive symptoms, a Glenn procedure (superior vena caval to right pulmonary artery anastomosis) may be life-saving but the risk is high.[47]

Severe Obstruction. The most common cause of death due to heart disease during the first week of life is hypoplasia of the left heart[35] (see Table 4). In this lesion the electrocardiogram shows right ventricular hypertrophy and the chest roentgenogram, pulmonary congestion. Patients with this condition all have pulmonary venous hypertension due to the absence of an adequate egress from the pulmonary veins or the left atrium or both, and they may also have systemic hypoperfusion due to an inadequate access route to the aorta or due to hypoplasia or aplasia of the ascending aorta and arch. When systemic hypoperfusion is present, the peripheral pulses usually cannot be felt and the flush blood pressure is below 40 mm of mercury.

Surprisingly, these babies usually appear normal at birth. This is, in part, owing to the fact that pulmonary flow and pulmonary venous pressure are low due to the persistence of the high fetal precapillary vascular resistance and to persistent patency of the ductus which provides an access route to the aorta from the right side of the heart. As the pulmonary vascular resistance falls, pulmonary blood flow, pulmonary venous volume and pulmonary venous pressure increase, resulting in a decreased pulmonary compliance and progressive dyspnea. Sudden vasoconstriction of the ductus may result in an acute "shock-like" clinical state which mimics the lethargic, ashen appearance of sepsis.

Most of these babies cannot be salvaged, but occasionally in those without systemic hypoperfusion, critical pulmonary venous hypertension can be relieved by atrial septectomy. In those with systemic hypoperfusion, when the aorta is of adequate size, there is experimental evidence to indicate that a Schumaker procedure[42] and distal banding of the pulmonary arteries may give effective palliation.[6] As there is no adequate egress from the left atrium, pulmonary venous hypertension is also

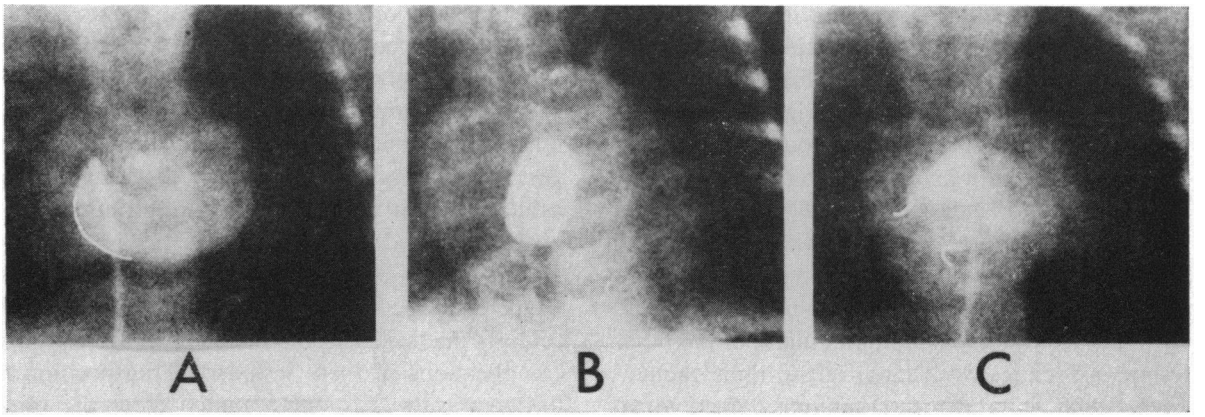

Figure 8.—Use of the Rashkind catheter allows for "non-surgical" creation of an atrial septal defect in patients with transposition of the great arteries. This procedure can usually be accomplished during diagnostic cardiac catheterization. The single frame from a cineangiogram on the left (A) illustrates the intact atrial septum during a left atrial cineangiogram. The middle frame (B) shows the balloon catheter inflated in the left atrium and the frame on the right (C) outlines the size of the atrial defect which was created after pulling the inflated balloon from the left atrium to the right atrium.

commonly the lethal physiological abnormality in patients with mitral atresia, as it is in patients with HLH, and an atrial septectomy may provide effective palliation.

Frequently single ventricle and transposition with or without pulmonary stenosis are associated with mitral atresia and the anatomical and clinical spectrum is broad. Aortic coarctation, characterized by strong axillary and absent or weak femoral pulses, is a common cause of distress after the first week of life (Table 4). When progressive failure occurs, cardiac catheterization is indicated to rule out complicating lesions. A preoperative aortogram is desirable for accurate identification of the location and length of the narrowed segment. Usually correction is possible by a relatively simple aortoplasty rather than resection and end-to-end anastomosis.[27] The clinical findings of critical uncomplicated valvar aortic stenosis are usually atypical during the neonatal period and there may be no systolic murmur. Observation of the negative contrast ejection stream from the left ventricle on the cineaortogram is a quick and effective way to establish this diagnosis.[7] Frequently there is complicating hypoplasia of the aortic annulus,[39] severely malformed valve leaflets or endocardial fibroelastosis,[23] and the salvage is consequently low. Aortic valve replacement has been successful in a few children but is technically very difficult in small infants.[43]

The problem of severe valvar pulmonary stenosis has been discussed previously under the cyanotic lesions. However, occasionally there are pronounced signs of right-sided failure when cyanosis is minimal or absent. Emergency valvotomy is lifesaving in such cases.[12] The hemodynamic handicap of patients with tricuspid atresia is occasionally accentuated by an inadequate egress from the right atrium. Creation of an atrial defect may be more important than changing the pulmonary blood flow by a shunting or banding procedure. The clinical picture of total anomalous pulmonary venous drainage below the diaphragm is completely different from drainage above the diaphragm, due to the fact that in the former there is pulmonary venous obstruction and hypertension. The heart is usually not enlarged and the chest roentgenogram shows pulmonary congestion.

Without operation these patients all die within six to eight weeks.[40] Total correction is difficult and dangerous. Experimental studies have shown that an atrial septectomy and distal pulmonary artery banding may provide effective palliation.[5]

Congenital mitral stenosis is rare and is usually associated with ductus and coarctation.[30] The diagnosis, even with cardiac catheterization and cineangiography, is difficult and must be differentiated from cor triatriatum and pulmonary venous obstruction. Surgical relief is rarely required during the neonatal period. Due to the pronounced deformity of the valve leaflets, relief usually requires replacement of the valve.

Primary Myocardial Disease (PMD). Endocardial fibroelastosis and myocarditis are the only common causes of primary myocardial disease during the neonatal period.[8] Accurate clinical differentiation of these two lesions is usually impossible. Both usually present a triad of massive cardiac enlargement (roentgenographically), progressive congestive failure, including gallop rhythm, and no significant murmur.

The electrocardiogram may show decided left ventricular hypertrophy and a "strain" pattern (Figure 9) suggesting endocardial fibroelastosis or there may be ST segment and T wave changes consistent with myocarditis.

Vigorous anticongestive therapy for both of these types of primary myocardial disease is indicated, but the salvage is low and persistent chronic failure is common.[28] Steroid therapy has been reported to be life-saving in some patients with myocarditis.[1]

Other forms of primary myocardial disease such as aberrant left coronary, glycogen storage disease, coronary vasculitis and idiopathic ventricular hypertrophy are rare and rarely cause symptoms during the neonatal period.

All patients with progressive distress from PMD should have angiographic visualization of the coronary arteries to rule out the surgically treatable lesion of aberrant left coronary and other potentially curable lesions which may mimic PMD. Congenital ventricular diverticulum is an extremely rare lesion which may mimic the clinical picture of PMD. The diagnosis can be made by palpation of the cardiac impulse in the epigastrium. Sudden rupture is common and salvage by early excision of the diverticulum is possible.[29] Acute distress from pericardial tamponade or constrictive pericardial disease is also rare in neonates but is usually occult and may masquerade as PMD. Recovery is possible if the disease is recognized and drainage and appropriate antimicrobial therapy instituted.[8,14]

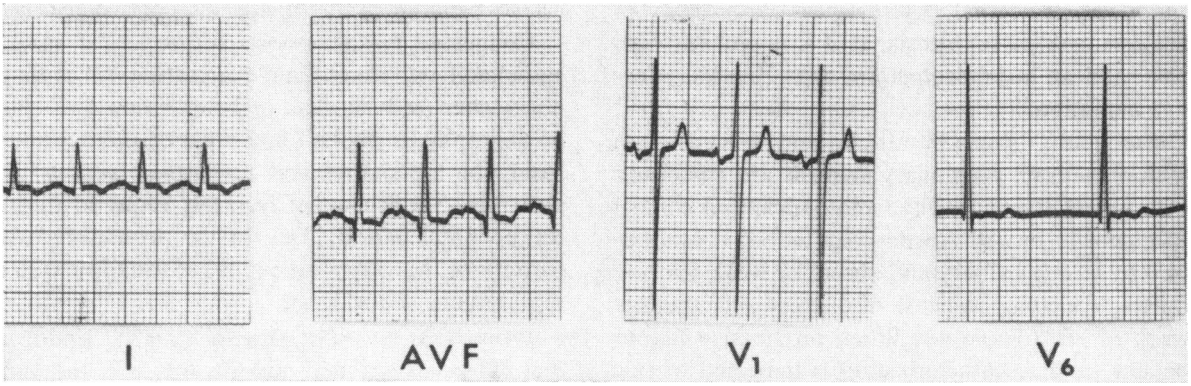

Figure 9.—The electrocardiogram, from a neonate who died of endocardial fibroelastosis, shows pronounced left ventricular hypertrophy and left ventricular "strain."

Massive Valvar Regurgitation. The regurgitant lesions are very rare and are included in this classification only for the sake of completeness. Isolated congenital valvar regurgitation is suspected from the unusual location (an apical systolic murmur with mitral insufficiency) or timing of the murmur (an early decrescendic diastolic murmur along the left sternal border with semilunar insufficiency) since apical murmurs and isolated semilunar insufficiency murmurs are infrequent in other types of neonatal cardiovascular disease.*

Patients with mitral insufficiency may have corrected transposition of the great vessels with Ebstein's anomaly of the left atrioventricular valve[41] or Marfan's syndrome. In the neonate, a loud aortic insufficiency murmur is usually due to a coronary fistula or a ruptured sinus of Valsalva rather than a regurgitant aortic valve.[18] Hemodynamically significant pulmonary insufficiency has also been reported in the newborn.[24] When there are progressive symptoms from a regurgitant lesion, surgical palliation by valvuloplasty or valve replacement is possible but the risk is high.[37]

Airway Obstruction. The neonate with a vascular ring most often first comes to medical attention because of a feeding problem, and formula changes are frequent before it is suspected that the stridor and dysphasia have a mechanical cause. The diagnosis is suspected on a routine chest roentgenogram by visualization of constriction of the negative tracheal shadow and by indentation of the esophagus following barium swallow. Cardiac catheterization and angiographic demonstration of the vessels involved are usually advisable (Figure 10). Correction should not be delayed, since a minor respiratory infection may suddenly accentuate the respiratory obstruction and result in sudden death. Following release of a vascular ring, stridor and symptoms similar to those of laryngotracheal malacia often persist for three to six months or longer. These residual symptoms may be related to persistent tracheobronchial changes which are secondary to the abnormal aerodynamics of the mechanical obstruction.

Arrhythmia. Paroxysmal atrial tachycardia is the most common arrhythmia during the neonatal period, and if it is not recognized death almost always ensues. The condition occurs mostly in boy babies and usually they do not have associated cardiac defects.[33] Reversion to sinus rhythm usually occurs within three to six hours following the beginning of digitalization. Occasionally reversion by electrical DC cardioversion is indicated.[36,45] Recur-

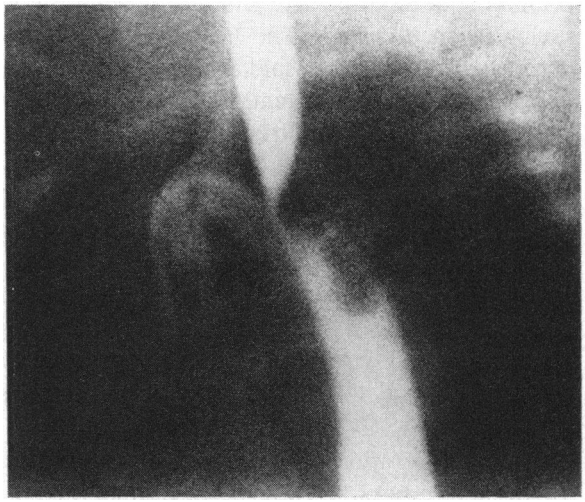

Figure 10.— Combined cineangiographic and barium swallow procedures demonstrate the vascular ring is due to a double aortic arch.

*The murmur of mitral insufficiency is heard in some patients with endocardial cushion defects and endocardial fibroelastosis. Semilunar insufficiency occurs in a small number of patients with ventricular septal defects when there is herniation of the non-coronary aortic cusp and in 10 to 25 percent of patients with truncus arteriosus.

rences are infrequent when maintenance digitalis is continued for six to twelve months. Ten to 20 percent of patients show WPW type conduction following reversion to sinus rhythm. Other forms of rapid heart beat, such as flutter and ventricular tachycardia, occur but are very rare. Analysis of the electrocardiogram is necessary for accurate differential diagnosis. The presence of apical clicks[21] and a variable intensity of the first sound[46] may be found in patients with ventricular tachycardia in contrast to those with supraventricular tachycardia.

Congenital complete heart block infrequently results in distress under two years of age unless there are associated cardiac anomalies causing significantly increased hemodynamic overloads. The first episode of cardiac arrest is rarely lethal and use of a pacemaker in children with uncomplicated heart block is not indicated until there have been two or more syncopal attacks. Heart block is more frequent in patients with corrected transposition of the great vessels.[41] Arrhythmias of all types are more frequent in patients with Ebstein's anomaly of the tricuspid valve.[32]

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